A Typical Presentation of Autoimmune Hemolytic Anemia with Stage 0 Chronic Lymphocytic Leukemia

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Introduction

- Autoimmune Hemolytic Anemia:
  - Most common autoimmune complication of CLL
  - Antibodies against someone's own RBC's causing them to lyse
  - Symptoms: Jaundice, weakness, dark urine, fatigue, dyspnea

Figure 1

Test	Expected Result in AIHA
Direct antiglobulin test (DAT)	Positive IgG ± C3 in warm-antibody–mediated AIHA C3 in cold-antibody–mediated AIHA
Haptoglobin	Decreased
Indirect bilirubin	Increased
Lactate dehydrogenase	Increased
Reticulocyte count	Increased
Urine hemosiderin	Present

Case Description

- 61-year-old female presenting to the ED with progressive dyspnea on exertion and fatigue
- Recently traveled to and from Idaho by car and attributed her symptoms to elevated altitude with hiking and travel fatigue, but symptoms were not improving
- PMHx: Stage 0 CLL, fallopian tube cancer s/p oophorectomy and chemo, childhood polio
- Stage 0 CLL diagnosed 9 months prior to arrival, 13q chromosomal deletion and IgVH mutation
  - Was under surveillance without treatment on arrival
- Patient was clearly jaundice but hemodynamically stable on arrival

Case Description

- Laboratory workup significant for:
  - Hgb: 5 gm/dL, WBC: 18,300 WBC/mm^3, reticulocytes: 19.47%, Total Bilirubin: 5.6 mg/dL, Direct Bilirubin: 0.2 mg/dL
  - Blood smear: No schistocytes, smudge cells appreciated
  - LDH: elevated at 500 U/L
  - Haptoglobin: <15 mg/dL
  - Direct Coombs Test: positive
  - Anti-C3 antibodies: positive
  - Anti-IgG Coombs antibodies: positive
  - Normal PT/PTT, fibrinogen
- Doppler US ruled out DVT to help rule out PE in setting of her recent travel and dyspnea on exertion
- Consented and transfused 2 units packed red blood cells
- Started on 40 mg IV Solumedrol BID
- Monitored for stabilization of her hemoglobin after transfusions and initiation to assure no evidence of ongoing hemolysis which was achieved by inpatient day 2
- Was transitioned to 1 mg/kg oral Prednisone dosing on discharge
- Followed up with hematology/oncology as an outpatient to be started on anti-CD20 therapy with rituximab

Discussion

- Autoimmune hemolytic anemia (AIHA) is the most common autoimmune complication of chronic lymphocytic leukemia (CLL)
  - Can present at any stage, often heralding the diagnosis
- Approximately 10% of patients with CLL experience AIHA with multifactorial and incompletely understood pathogenesis
- Regardless of CLL staging, if associated with autoimmune hemolytic anemia, treatment becomes indicated
- Before initiating steroid treatment, DIC and TTP need to be excluded as differential diagnoses as their treatment differs and a missed diagnosis could be fatal
  - Would see schistocytes on blood smear and abnormalities in PT/PTT and/or fibrinogen

Conclusion

The differential diagnosis for progressive anemia in patients' with CLL must include AIHA, and de novo AIHA should prompt investigation for possible early stage CLL.

References

Figure 1.
Tests for Autoimmune Hemolytic Anemia. [Online Image]. (Oct 2019). Retrieved April 16, 2023 online